

Case Report

Good visual outcome in an immunocompromised patient with bilateral acute retinal necrosis syndrome A case report



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Abstract

Acute retinal necrosis (ARN) is an uncommon necrotizing, fulminant retinopathy caused by the *herpes simplex* virus types 1 or 2 or by the *varicella zoster* virus with visually devastating consequences. Generally it occurs in patients who are systemically healthy, but occasionally occurs in immunocompromised host. We report a case of bilateral ARN in a patient with AIDS with a good final visual outcome.

Keywords: Acquired immunodeficiency syndrome, Herpes simplex, Retinal necrosis syndrome, Acute, Retinal detachment, Uveitis

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<http://dx.doi.org/10.1016/j.sjopt.2013.11.005>

Introduction

Acute retinal necrosis (ARN) syndrome is an uncommon but devastating potentially blinding necrotizing retinitis. Characteristically presents as a peripheral retinal vasculitis. It can affect one or both eyes. It is observed most commonly in healthy patient but occasionally occurs in immunocompromised host.^{1,2} Most, if not all, cases of ARN syndrome are caused by infection with a member of the herpes virus family, usually *varicella zoster* virus (VZV) or *herpes simplex* virus (HSV) type 1 and more rarely by HSV-2 or *cytomegalovirus* (CMV).³

The disease is characterized by anterior uveitis, vitritis and patchy or confluent areas of white- or cream-colored areas of necrotizing retinitis that rapidly extend posteriorly from the peripheral retina. Retinal atrophy resulting from necrosis often leads to secondary rhegmatogenous retinal detachment (RD). Occlusive vasculitis involves the retinal and choroidal vasculature.⁴

Current treatment trends vary widely, including single agents or combination of oral, intravenous, and intravitreal agents. Differing strategies do not affect outcomes. The final visual acuity in ARN is generally poor. RD is common and can neither be predicted nor prevented.

Case report

A 30-year-old white male, previously immunocompromised with AIDS, presented with a 3-week history with blurred vision, floaters and decreased peripheral vision in both eyes. He has been under treatment for AIDS for 10 years. His medications included abacavir, lamivudine, lopinavir, ritonavir, itraconazole and ganciclovir. His CD4 cell count was 66/mm³ and viral load was 75.589/mm³.

On examination, the patient's best visual acuity (BVA) was 20/50 OU. The intraocular pressure was 14 mmHg and 15 mmHg in the right and left eye, respectively. Fine keratic precipitates, trace anterior chamber cells, 1+ anterior

Received 22 June 2013; received in revised form 12 October 2013; accepted 8 November 2013; available online 18 November 2013.

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Peer review under responsibility of Saudi Ophthalmological Society, King Saud University



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vitreous cells was present in both eyes. Dilated fundus examination showed retinal arteriolar sheathings in all quadrants, extensive areas of whitening and retinal necrosis involving the retina periphery in both eyes (Fig. 1). A diagnosis of bilateral ARN was made, the ganciclovir was changed for intravenous aciclovir for 10 days, and he was prescribed topical corticosteroids and atropina. One week after the initial presentation prophylactic laser retinopexy was performed, completely demarcating involved areas in both eyes (Fig. 2). In the next two weeks, there was a dramatic resolution of retinal necrosis resulting in areas of retinal atrophy, his BVA has improved to 20/30 OU. Patient returned one week later with complaint about his vision, his BVA was 20/100 in the right eye and 20/80 in the left eye. On fundus examination retinal detachment with macula involvement was noted in both eyes. The patient underwent pars plana vitrectomy, endolaser treatment, and silicone oil tamponade. The left eye had a redetachment, which was surgically repaired in the same way two weeks later (Fig. 3).

After two months of the initial presentation his BVA was 20/30 in the right eye and 20/200 in the left. His medication was changed to oral aciclovir 800 mg 5 times daily for 14 weeks. 5 months after the last vitrectomy vision in both eyes remains stable.

Discussion

Acute retinal necrosis syndrome is an infectious uveitis usually caused by HSV-1 or -2 or VZV. Features of ARN syndrome include a vaso occlusive angiitis of both retinal and choroidal vessels, a necrotizing retinitis that

preferentially involves the peripheral retina, and significant intraocular inflammation. Rhegmatogenous retinal detachment is a major late sequela. Urayama et al. first described ARN syndrome in 1971.⁴ It has a bimodal age distribution peaking at approximately 20 and 50 years of age. There is no clear racial association.⁶ Although the syndrome was initially described in healthy patients, it subsequently has been reported in individuals with compromised systemic immunity, most notably in those with human immunodeficiency virus (HIV) infection.⁷ The ARN syndrome was seen at any level of HIV infection.⁸ The ARN syndrome can be insidious or present with floaters, blurred vision, or rarely decreased peripheral vision. External examination may reveal diffused episcleritis, scleritis, or orbital inflammatory disease. Mild to moderate anterior segment cellular reaction is common. ARN syndrome is characterized by a retinal vasculitis affecting both the arteries and veins in the fundus, which is manifested by sheathing of the major vessel. Peripheral retinal whitening is typically present, and the entire peripheral retina may be involved. Vitritis and optic disk swelling either hyperemic or pallid, are common features of the ARN syndrome. The retinitis may not progress posteriorly to the vascular arcades, sparing the macula and central vision. Without treatment, the inflammatory component of ARN syndrome typically burns out, leaving behind a thin atrophic retina with pigmentary changes. Rhegmatogenous retinal detachment is a major cause of visual loss and occurs in 50–75% of cases. Vitreous traction and proliferative vitreoretinopathy are complicating factors.⁷

The diagnosis of ARN syndrome is based on a clinical examination and a characteristic fundoscopic appearance.

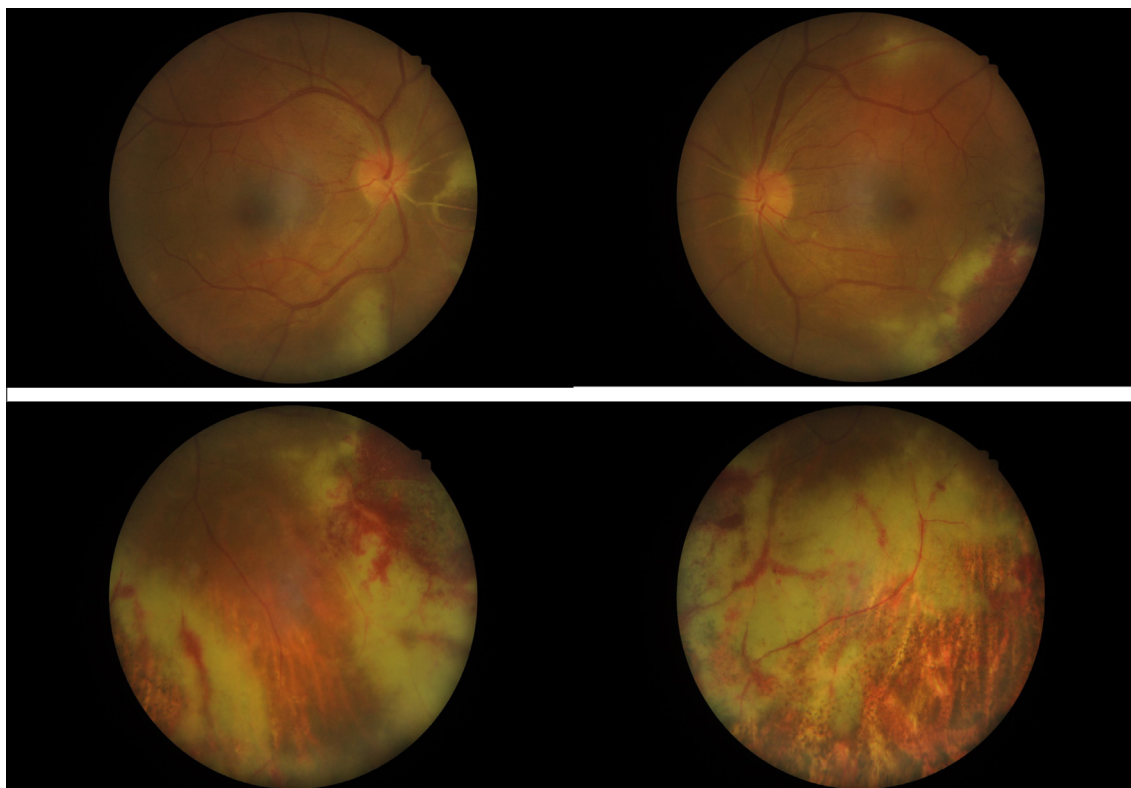


Figure 1. The arteriolar sheathings in all four quadrants, extensive areas of whitening and retinal necrosis involving the retina periphery for 360° in both eyes.

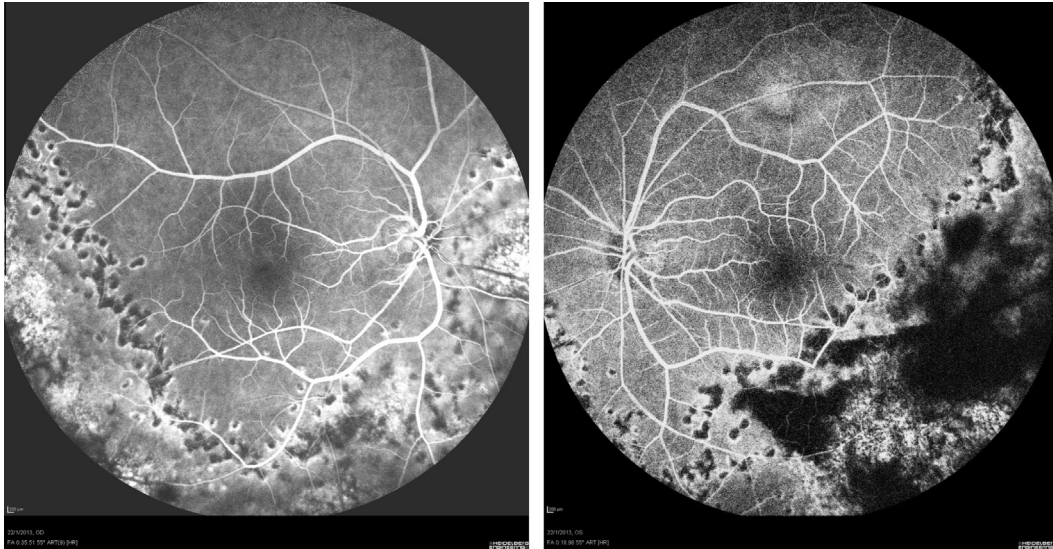


Figure 2. Prophylactic laser retinopexy.

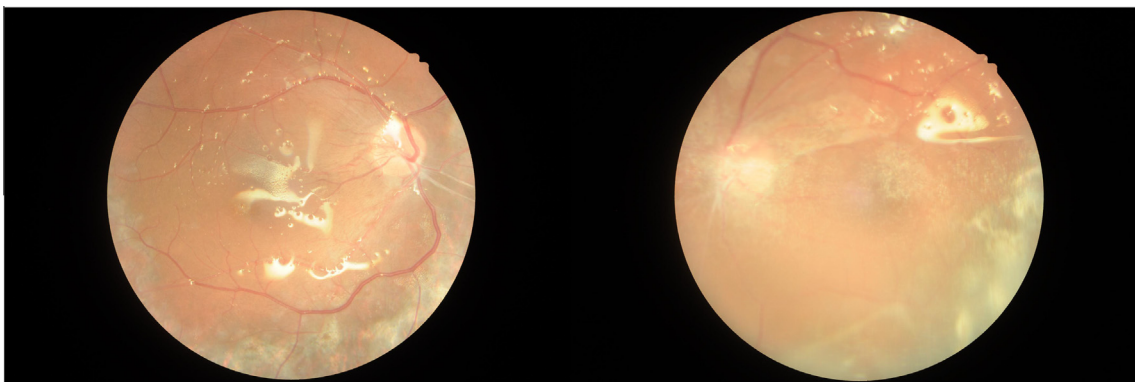


Figure 3. After the pars plana vitrectomy with silicone oil tamponade in both eyes.

Diagnostic vitrectomy or retinal biopsy may be indicated in some atypical cases. Differential diagnosis included cytomegalovirus retinitis and progressive outer retinal necrosis (PORN).

Intravenous acyclovir is the current medical treatment of choice for active ARN syndrome. Initial treatment with a 10 days course of high dose intravenous acyclovir (10 mg/kg every 8 h) is followed by oral acyclovir (800 mg taken 5 times a day) for up to 14 weeks. The long-term oral maintenance therapy may be required in immunosuppressed patients who develop recurrent lesions. Ganciclovir and foscarnet are alternative intravenous agents. Intravitreal injections of ganciclovir or foscarnet may be considered to treat individuals limited by systemic drug toxicity.⁷ Anticoagulation to treat the vascular obstructive component of the ARN syndrome has been recommended by some investigators.⁵⁻⁷ Systemic corticosteroids help to control and limit the damage caused by the severe inflammation associated with ARN syndrome. Prophylactic laser barrier treatment is recommended at the earliest opportunity preferably within the first 2 weeks to reduce the incidence of RD.¹

Vitrectomy with silicone oil tamponade is mandatory to achieve retina reattachment for rhegmatogenous and

traction retinal detachment with multiple retinal tears after the fulminant type of ARN syndrome.⁹

Sternberg et al. noted RD in 17% of the eyes treated prophylactically with laser compared with 67% of untreated eyes. Earlier previous reports indicated that only 23% of the detached retinas are successfully reattached, and only 28% of the eyes affected by ARN syndrome eventually achieved visual acuities of 20/200 or better.⁷⁻¹⁰ Our patient achieved the best visual acuity of OD 20/30 and OS 20/200.

The good visual results may be explained by the prompt diagnosis, proper and early treatment of the complications. Hillenkamp et al. hypothesized that an intensified therapeutic approach including early vitrectomy with intravitreal acyclovir lavage, and, when feasible, laser demarcation of necrotic retinal areas, scleral buckling, and gas or silicon oil tamponade, may help to halt the progression of the disease, to prevent secondary RD and phthisis bulbi, and to help to preserve visual function.⁴ Because the poor prognosis associated with retinal detachment, prophylactic treatment remains an important issue.¹⁰ The timing of surgical intervention is crucial to good visual outcome in prophylactic and early vitrectomy for acute retinal necrosis.⁹ Prospective randomized

clinical trials are need to evaluate the efficacy of the treatment in bilateral ARN syndrome.

Conflict of interest

The authors declared that there is no conflict of interest.

Financial disclosure(s)

The authors have no proprietary or commercial interest in any materials discussed in this article.

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